

Improvement in Pulmonary Function After Closure of Atrial Septal Defect in a Patient With Cystic Fibrosis

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Abstract

Atrial septal defect (ASD) is a major cause of left-to-right intracardiac shunting. If persisting into adulthood, an ASD can lead to a larger shunt, which may eventually cause pulmonary hypertension and right ventricular failure. Large intracardiac shunts cannot be tolerated in patients with underlying lung disease such as cystic fibrosis. Although the association between an intracardiac shunt and cystic fibrosis has been reported in the literature, the impact of ASD closure on the clinical course of patients with cystic fibrosis has not been studied. Here, we report a case of ASD closure in a patient with cystic fibrosis with hypoxemia out of proportion to his lung disease. Closure of the ASD shunt resulted in significant improvement of his symptoms and pulmonary function testing.

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Key Words

Atrial septal defect • Septal closure device • Cystic fibrosis

Introduction

Secundum atrial septal defect (ASD) is an isolated defect in the fossa ovalis between the atria. Although many ASDs close spontaneously during the first year after birth [1], the defect persists in some cases, causing a left-to-right shunt [2]. Left-to-right shunting,

when significant, can cause dyspnea on exertion, which may not be well tolerated in patients with an underlying pulmonary condition such as cystic fibrosis. It is unknown whether ASD closure could lead to symptomatic improvement in these patients. Here, we report a case of ASD closure resulting in improved pulmonary function testing in a patient with cystic fibrosis with a bidirectional shunt and worsening functional capacity.

Case Presentation

Our patient was a 36-year-old man with cystic fibrosis diagnosed at birth with a dF508/dF508 genotype. His symptoms could be characterized as moderate airway disease complicated by chronic airway infections, pancreatic insufficiency, diabetes, and malnutrition. Progression of his clinical condition caused dyspnea with minimal exertion. He was referred to our cardiology service to be evaluated for hypoxemia out of proportion to his chronic pulmonary disease. Pulmonary function testing showed a forced expiratory volume in 1 s (FEV1) of 1.71 (43% predicted). Transesophageal echocardiography (TEE) showed a small ASD with a bidirectional shunt seen at rest (Figure 1). Right heart catheterization showed a pulmonary artery pressure of 26/7/16 mmHg and pulmonary vascular resistance of 1.7 Wood units. A decision was made to proceed with ASD closure, which



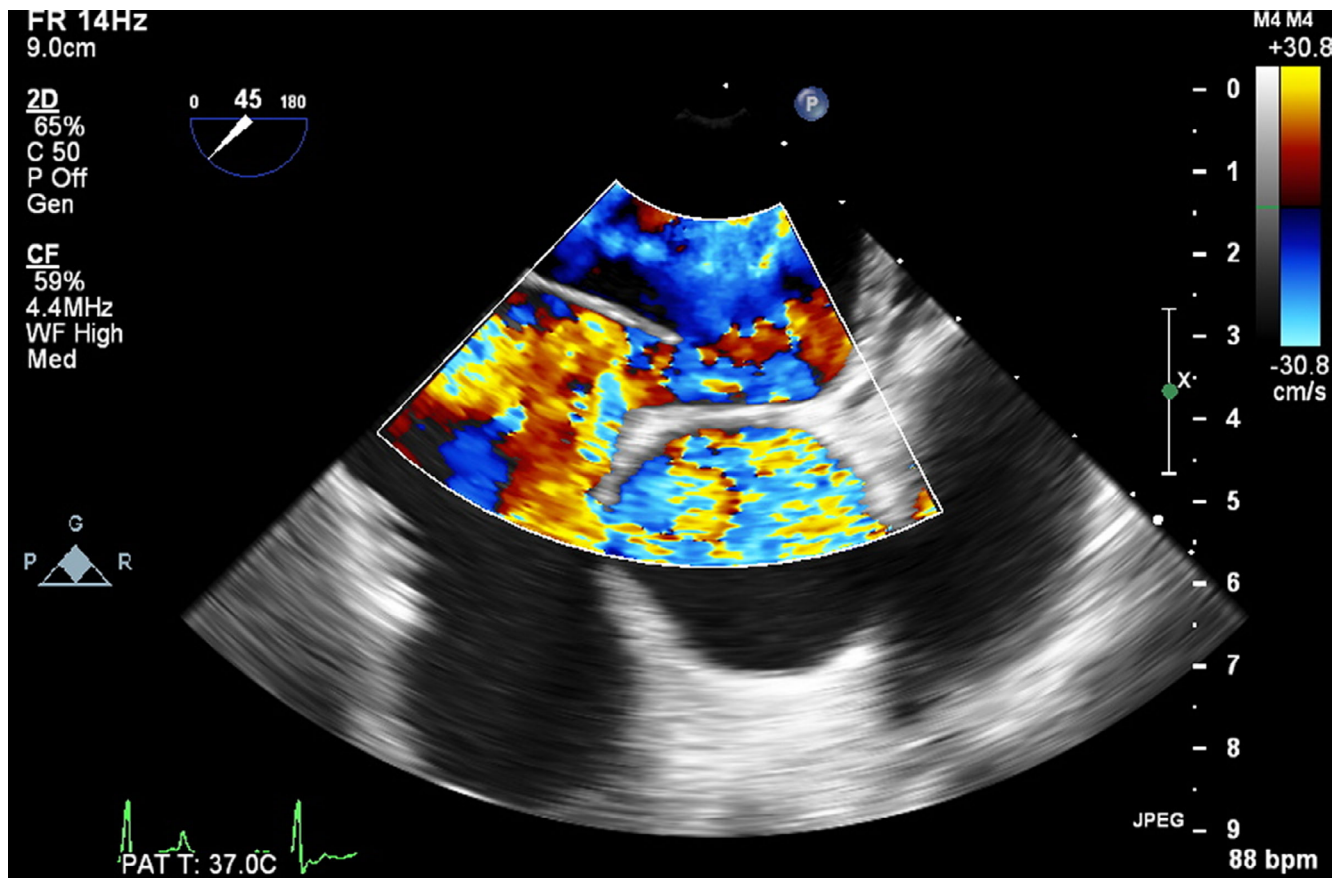


Figure 1. TEE showing bidirectional shunt before 20-mm Occluder Septal Helix placement.

was performed under the guidance of intra-cardiac echocardiography and fluoroscopy. A balloon sizing the defect was used, and the patient underwent ASD closure with a 25-mm Occluder Septal Helix. The indication for this closure was the presence of bidirectional flow that would ultimately lead to irreversible pulmonary hypertension. At the end of the procedure, an agitated saline study was performed, which revealed minimal residual shunt. Echocardiography performed the next day showed very minimal flow across the device as demonstrated by color Doppler. The patient was discharged to home on 81 mg aspirin and 75 mg clopidogrel daily for 3 months. He initially demonstrated symptomatic improvement with less oxygen requirement at rest. However, 3 months later, his symptoms of fatigue and dyspnea on exertion returned, and follow-up echocardiography demonstrated a large right-to-left intra-cardiac shunt at rest as demonstrated by an agitated saline study

(Figure 2). His pulmonary function testing during this time showed an FEV1 of 1.78 (47% predicted). The patient was monitored for 1 year, during which his functional capacity and cystic fibrosis symptoms remained stable. He subsequently suffered from a neurological event suggestive of a transient ischemic attack of an embolic nature, after which he was considered for residual shunt closure. TEE revealed a deformity of the Helix device causing a bidirectional shunt via the inferior rim of the device. With further questioning, the patient admitted to using a high-frequency chest wall oscillation device a few days after his ASD closure as part of his routine treatment for cystic fibrosis.

The patient underwent closure of the residual shunt using a 30-mm Cardioform device. The second device was advanced across the defect and positioned to sandwich both sides of the previously placed Helix Occluder (Figure 3). The absence of a

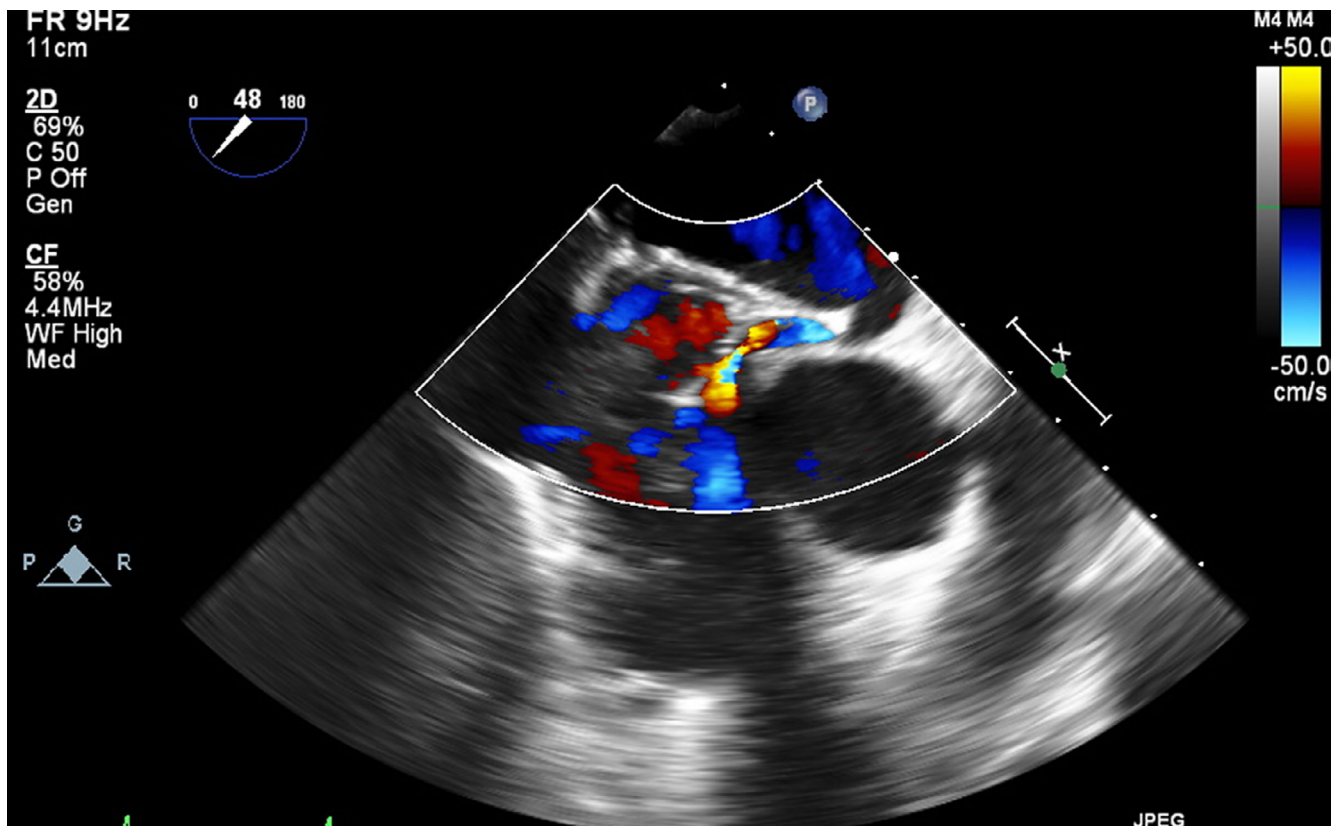


Figure 2. TEE showing residual shunt 1 year after 20-mm Occluder Septal Helix implantation.

residual shunt was confirmed with several negative agitated saline injections. He was advised to not use the high-frequency chest wall oscillation device for at least 12 months. At 6-month follow-up, the patient showed significant improvement in his functional capacity, with pulmonary function testing demonstrating a spike increase in FEV1 to 3.4 (89% predicted; Table 1).

Discussion

ASD is one of the most common adult congenital heart defects [3, 4]. It is caused by underdevelopment of the secundum septum or over-reabsorption of the primum septum. In most cases, ASDs close spontaneously during infancy [4, 5]; however, if persistent, their clinical impact is related to their location, size, and association with other defects [5, 6]. Small ASDs usually cause a left-to-right shunt without significant structural consequences in the right-sided heart chambers. However, larger ASDs, if not corrected in

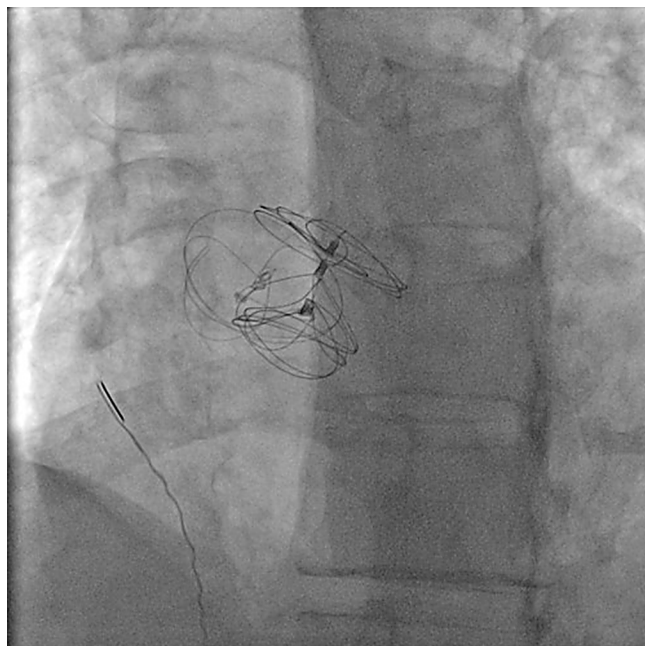


Figure 3. Fluoroscopy showing placement of a 30-mm Cardioform device adjacent to the previously placed 20-mm Occluder Septal Helix.

Table 1. Pulmonary function test of the patient over his clinical course.

	Before first ASD closure	After first ASD closure (with residual shunt)	After second ASD closure (no significant residual shunt)
FEV1	1.71 (43% predicted)	1.78 (47% predicted)	3.4 (89% predicted)
FEV1/FVC	53 (65% predicted)	61 (76% predicted)	76 (95% predicted)

early life, may lead to right-sided volume followed by pressure overload [6, 7], resulting in right atrial and ventricle enlargement and ultimately pulmonary vasculature remodeling and pulmonary hypertension with symptoms of right heart failure such as fatigue, decrease in functional capacity, and lung infections [7, 8]. The association between ASDs and cystic fibrosis is not well established. Two small series of patients with cystic fibrosis reported a 53–55% incidence of left-to-right shunt, mainly due to the presence of a patent foramen ovale (PFO) as seen by TEE [8, 9]. We can assume that patients with severe lung disease, such as cystic fibrosis in our case, will not tolerate hypoxemia caused by a significant right-to-left shunt. Current American College of Cardiology/American Heart Association guidelines [10] recommend percutaneous interventional therapy for ASDs in the presence of right atrial and right ventricle enlargement with or without symptoms before progressing to irreversible pulmonary hypertension, the presence of which is considered a contraindication for ASD closure. The impact of ASD closure on pulmonary function testing in patients with cystic fibrosis is not known. Belton et al. [11] reported a 29-year-old man with cystic fibrosis who underwent PFO closure after developing neurological symptoms. His pulmonary function had deteriorated at the time of the neurological event. Closure of his PFO improved his pulmonary function with an increase in FEV1 from 1.36 to 1.89 (33% to 47% predicted). The authors concluded

that closure of the PFO may have had a significant impact on lung function. The indication for the first ASD closure in our case was the presence of bidirectional flow, for which we aimed to prevent the development of irreversible pulmonary hypertension that would preclude ASD closure. The first closure significantly improved pulmonary function testing before the device migrated and led to a residual shunt with worsening symptoms and test results. We assume that the use of a high-frequency chest wall oscillation device dislodged the initial occluder, resulting in larger residual shunt. The indication for the second closure was a neurological event caused by a paradoxical embolism. There was significant improvement in pulmonary function testing after the second ASD closure, which was not associated with a residual shunt immediately after the procedure.

In conclusion, percutaneous closure of ASD in patients with cystic fibrosis and hypoxemia out of proportion to their lung disease may improve pulmonary function testing and hypoxemia, although further studies are warranted to confirm this possibility.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

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